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Myeloid sarcoma as a manifestation of acute myeloid leukemia

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ABSTRACT

We report a case of a 43-year-old man who presented with dyspnea because of large bilateral pleural effusions and imaging findings of a large periaortic mass with compression of the esophagus and left atrium. Subsequent soft tissue biopsy was consistent with myeloid sarcoma, and bone marrow biopsy was consistent with acute myeloid leukemia. He was started on induction and subsequent consolidation chemotherapy with complete remission and shrinkage of the tumor.

KEYWORDS Acute; extramedullary manifestations of hematological malignancies; myeloid sarcoma

Soft tissue masses are a rare presentation of hematological malignancies, including acute myeloid leukemia (AML), myeloproliferative disorders, and myelodysplastic syndrome.[1,2] Historically, these tumors have been referred to as "chloromas" due to their high content of myeloperoxidase, which gives these tumors a green color.[1,2] Myeloid sarcoma can arise in a variety of body tissue sites, including bone, gastrointestinal tract, peritoneum, lymph nodes, and skin and soft tissue.[1]

CASE PRESENTATION

A 43-year-old man with a known history of tobacco use, posttraumatic stress disorder, hyperlipidemia, and major depressive disorder presented to the emergency department with progressively worsening orthopnea, substernal chest pain, nonproductive cough, and palpitations. He had a 30 pack-year smoking history, social alcohol use, and daily marijuana use. Significant findings included a D-dimer of 5.62 μg/mL, elevated C-reactive protein of 8.7 mg/dL, and an erythrocyte sedimentation rate of 54 mm/h. Other laboratory tests were normal, including complete blood count (no blasts on peripheral blood), comprehensive metabolic panel, brain natriuretic peptide, blood culture, troponin, IgE, IgG4, complements C3 and C4, hepatitis, human immunodeficiency virus, rheumatoid factor, autoimmune marker panel, antineutrophil cytoplasmic antibodies, myeloperoxidase, proteinase 3, and tests for endemic fungal infections.

A chest radiograph showed a large right- and left-sided pleural effusion. Subsequent computed tomography (CT) pulmonary angiography showed a soft tissue mass surrounding the descending thoracic aorta measuring 9.6 × 6.0 × 17.8 cm with mass effect on the esophagus and left atrium *(Figure 1)*. A transthoracic echocardiogram showed a left ventricular ejection fraction of 50%.

A bilateral thoracentesis showed an exudative effusion with atypical cells consistent with malignancy. Pleural fluid analysis showed immature myeloid (leukemic) cells with coexpression of CD34, CD117, HLA-DR, MPO, CD38, CD15, CD64, TdT (dim, partial), and CD19. Bone marrow biopsy flow cytometry showed a distinct population of immature myeloid cells representing 4% of the total cell population. Chromosome analysis using fluorescent in situ hybridization was remarkable for a variant fusion pattern of RUNX1T1 and RUNX1. An interventional radiology–guided biopsy of the mass showed a final pathology of myeloid sarcoma.

The patient was initiated on 7 + 3 induction chemotherapy with cytarabine followed by daunorubicin followed by consolidation chemotherapy with high-dose cytarabine. Subsequent surveillance CT scan and positron emission tomography of the chest showed shrinkage of the mediastinal

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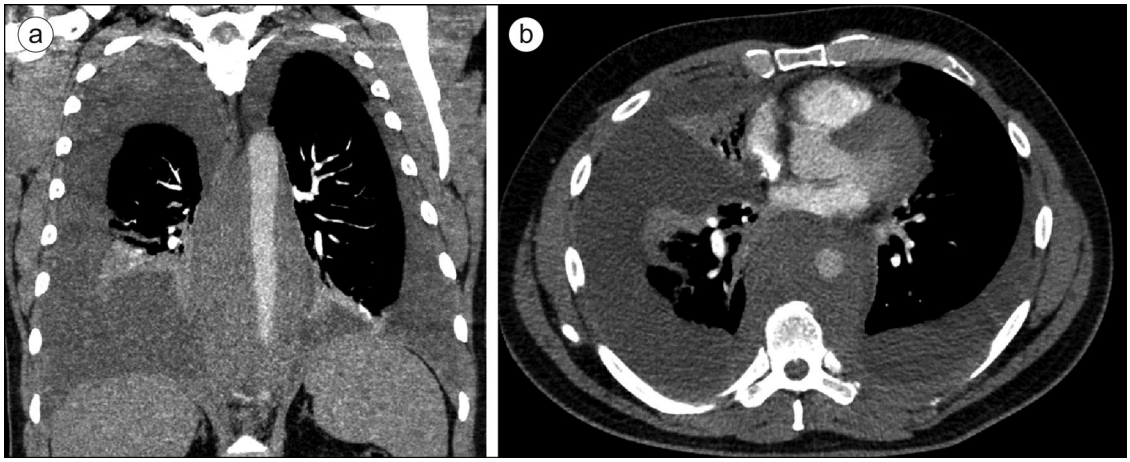

Figure 1. **(a)** Coronal and **(b)** axial CT revealing a soft tissue mass surrounding the descending thoracic aorta measuring 9.6 cm transverse × 6.0 cm anterior-posterior × 17.8 cm circumference with a mass effect on the esophagus and left atrium.

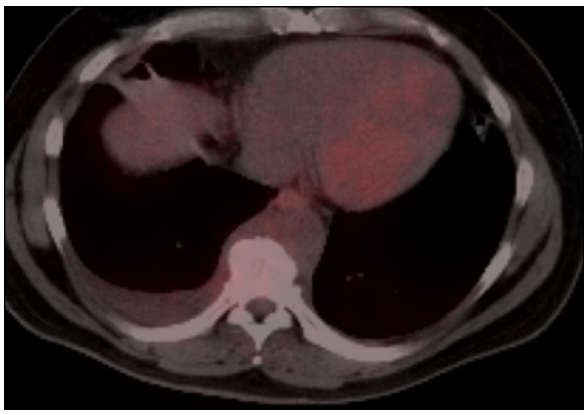

Figure 2. Positron emission tomography after initiation of chemotherapy showing interval decrease in size of the posterior mediastinal mass and no hypermetabolic findings to suggest active malignancy.

mass from 8.7 to 6.8 cm (largest axial dimension). He is currently in remission and tolerating chemotherapy *(Figure 2)*.

DISCUSSION

Myeloid sarcoma can manifest as the first clinical sign of AML, often appearing months to years before detectable atypical malignant cells or as a manifestation of relapsed AML after allogenic hematopoietic stem cell transplant.[1,3] Myeloid sarcoma has a minimal male predominance (1.2:1) and presents in 2.5% to 9.1% of AML cases.[1,3] The development of myeloid sarcoma, whether preceding AML or in relapsed treated AML, is poorly understood but theorized to involve cytokine receptors and adhesion molecules to specific extramedullary sites.[1]

The diagnosis of myeloid sarcoma is often a significant challenge, as it is asymptomatic until the patient experiences a mass effect from the tumor on surrounding structures.[1,2] Our patient developed dyspnea due to bilateral pleural effusions because of the mass effect on the aorta, esophagus, and heart. Imaging including CT and magnetic resonance imaging are important diagnostic and prognostic tools for myeloid sarcoma.[1,3] Core biopsy is often necessary for tissue diagnosis.[1,3] In many cases of primary myeloid sarcoma (presence of a soft tissue mass in the absence of the diagnosis of AML), misdiagnosis is common—historically up to 75%, and with recent advancements in immunohistochemistry, flow cytometry, and fluorescence in situ hybridization, up to 25% to 47%.[1]

Treatment is focused on current AML protocols, as myeloid sarcoma is usually found in conjunction with AML and most primary myeloid sarcomas will eventually progress to AML.[1,3,4] Local treatment with radiotherapy, surgery, or both can be considered following response to chemotherapy, following hematopoietic stem cell transplant, or as part of debulking.[1,3,4]

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